THE COSTS OF A CASE OF VARIANT CREUTZFELDT-JAKOB DISEASE

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Preventing a single case of variant Creutzfeldt-Jakob disease would generate large benefits. In what follows we describe the progress of a single case and provide an estimate of the value of preventing it.

While the classical form of CJD manifests itself in people between the ages of 55 and 75, with an average of 65 years, vCJD strikes a much younger population. The age of onset in vCJD patients has mostly been 18 to 50 years (about 9% under 18 and about 5% over 50). The average age of onset is 29 years. Research so far seems to indicate the vCJD has a rather long incubation period. How long the incubation period actually is has been debated, but can be as little as 5 years, and some researchers believe the maximum could be as long as 60 years. Currently there is no way to detect, treat, or cure vCJD; from disease onset until death, victims have a life expectancy of about 14 months.

Variant Creutzfeldt-Jakob disease is difficult to diagnose because the patient initially experiences non-specific symptoms. These non-specific symptoms may include depression, personality changes, apathy, anxiety, and painful or cold limbs. Within the first month or two of disease onset, the patient will likely visit a general practitioner. As the vCJD progresses, non-specific symptoms develop into true psychiatric symptoms that include depression, personality changes, withdrawal, delusions, and hallucinations. At this point in the progression of the disease, the patient will likely seek specialized medical care, including psychiatric evaluation. Many vCJD patients are initially misdiagnosed as having psychiatric illness, delaying the true diagnosis. Our vCJD cost estimates take into account the difficulty in diagnosing the disease by including the costs of various misdiagnoses and types of physicians seen.

After about 6 months, disease symptoms progress and vCJD patients begin to exhibit neurological as well as psychiatric symptoms. Patients experience involuntary movement, suffer memory loss, lose their ability to work, and need constant supervision. At the first signs of neurological symptoms, vCJD patients will begin seeing a

neurologist and are often admitted to hospital neurological units. The psychiatric symptoms of the vCJD patient do not abate with the coming of neurological problems; indeed, psychiatric symptoms become enhanced as patients can become aggressive or psychotic. Hospital staff in both psychiatric and neurological acute care units is often unprepared to deal with vCJD patients needing serious long-term care. Our vCJD cost estimates account for visits to a neurologist and inpatient hospital stays in a neurology ward.

The onset of neurological symptoms in vCJD patients marks the beginning of a rapid health decline ending in death. The worsening neurological symptoms cause the vCJD patient to become immobile, mute, and totally dependent on others for care. Hospital staff is often unable to keep up with the deterioration of the patient's condition, not realizing that the patient is no longer able to swallow food or use the bathroom. On average, variant Creutzfeldt-Jakob disease sufferers go from developing unsteadiness to becoming bed bound in 6 months and from becoming bed bound to death about 8 months later.

In addition to seeing a general practitioner, psychiatrist, and a neurologist, it is quite likely that a patient with vCJD will also receive palliative care for symptom and pain management, physical therapy and occupational therapy to slow patient coordination loss, and speech therapy to delay the onset of mutism. Often a dietician is consulted to make sure the vCJD patient gets proper nutrition. Eventually, the rapidly deteriorating vCJD victim will have to rely on PEG feeding (Percutaneous endoscopic gastrostomy-tube feeding) for their nutrition needs. The costs of palliative care, physical and occupational therapy, and tube feeding are taken into account in our estimates.

Physicians who are trying to diagnose a vCJD patient use many procedures. Patients can expect to undergo several MRIs, EEGs and CT scans, especially as neurological symptoms begin to appear. The purpose of these diagnostic tools is to examine the brain for any unusual occurrences, but evidence of vCJD often fails to show up on these scans. Scanning procedures often had to be repeated for other reasons, such as psychiatric symptoms, outbursts, and the patient's fear. After exhausting scanning procedures, if vCJD is the suspected illness, it is likely that a brain

biopsy or a tonsil biopsy will be performed. Brain biopsies are most likely to be performed post-mortem to confirm the cause of illness; however, brain biopsies can also be performed while the patient is still living. Besides affecting the brain and the rest of the nervous system, vCJD has been shown to affect the lymphoid system, so evidence of vCJD can show up in a biopsy of tonsil tissue. The MRI and EEG brain scans and biopsies are included in our disease cost estimates.

Care for a new variant Creutzfeldt-Jakob disease sufferer is expensive and time-consuming. Psychiatric counseling is needed for patient mental disturbances and physical therapy is needed to minimize the patient's neurological breakdown. Patients' needs change rapidly with the deterioration of their condition. Beyond physical therapy, neurological deterioration requires that the patient receive around-the-clock care, including tube-feeding and incontinence help.

Families of vCJD patients may chose to care for the patient at home once the permanency and finality of the illness is realized. Home care is mentally, physically and financially draining for the families. Patients' families often need counseling to cope with the rapidly deteriorating effects variant Creutzfeldt-Jakob disease has on their family member. Families caring for a vCJD patient at home also need financial assistance to cover lost earnings and pay for assistance with patients, household chores, childcare, and transportation. Medical professionals sometimes need counseling to deal with the emotional stress of caring for a vCJD patient. Our cost estimates do not take into account the secondary effects of vCJD on the patient's family and medical professionals.

If the family cares for the vCJD patient at home, health aids must be purchased. It is not uncommon for the vCJD-afflicted household to need the services of a home caregiver and such implements as a walker or a wheelchair, pressure mattress, hospital bed, and special toilet accommodations. If the family opts for in-patient hospital or hospice care they will not need to purchase home health aids but transportation, hospital parking and hospital cafeteria meals for family members become expensive. The costs of home health aids and the costs of both inpatient and outpatient hospice care are accounted for in our vCJD estimates.

There are other special problems that must be dealt with by vCJD patients' families. First, the media attention surrounding bovine spongiform encephalopathy causes the human sufferers of vCJD to gain unwanted media attention. Second, the anxiety and fear associated with BSE and variant Creutzfeldt-Jakob disease means that some caregivers may be hesitant to deal with a vCJD patient. Similarly, unfounded fear of contamination from vCJD patients also affects the families' ability to find a mortician to handle the patient's body after death. This problem often translates into higher funeral and burying costs for these vCJD patient families. Finally, there has been a feeling among vCJD patient families in Great Britain that the government is trying to "cover-up" or hide vCJD victims. The emotional costs of dealing with the media and higher funeral costs are not accounted for in our estimates.

The estimates presented below attempt to capture the full direct cost of an average case of vCJD. We apply costs from the United States to data on the disease progression, and treatment, of victims from the United Kingdom. Variant Creutzfeldt-Jakob disease differs greatly from other conditions. However, in some aspects, vCJD is similar to other debilitating, costly, and fatal conditions such as certain types of cancer, Alzheimer's disease, multiple sclerosis, amyotrophic lateral sclerosis (ALS-Lou Gehrig's disease), sporadic CJD, and AIDS. Wherever possible, we used cost data from known treatments of these diseases to help estimate the costs of a case of vCJD.

Table 1 shows the components of the costs per case of vCJD when a life is valued at $5 million (VSL) and the qaly loss for illness is $100,000 per year.

Table 1- Model of vCJD Costs per case for $5M + $100K*Qaly Loss

Baseline Values	
Length of stage 1 (months)	4
Length of stage 2 (months)	3
Length of stage 3 (months)	4
Length of stage 4 (months)	2

Length of illness (months)	13
$ per year	100,000

Medical costs	
stage 1	$29,329
Mean Number of visits for medical care in stage 1	2
Total cost stage 1	$58,657
stage 2	$11,700
Mean Number of visits for medical care in stage 2	6
Total cost stage 2	$70,201
stage 3	
Hospice care outpatient costs	$19,901
health aids (all stages)	$11,611
Total cost stage 3	$31,512
stage 4	
Hospice care inpatient costs	$29,471
Total cost stage 4	$92,495
Total Costs All Stages	$252,865
procedures (all stages)	$370,572
drug costs (all stages)	$16,344
Total $ medical	**$639,782**

Lost utility	
stage 1 qaly loss	0.318
stage 2 qaly loss	0.523
stage 3 qaly loss	0.585
stage 4 qaly loss	0.613
years in stage 1	0.30769
years in stage 2	0.23077
years in stage 3	0.30769
years in stage 4	0.15385
utility lost (in QALY)	0.49285
$ per year	$100,000
Total $ lost utility	**$49,285**

Death-Value of a Statistical Life	$5,000,000

Results	
Total costs per case	$5,689,066

The cost of a case of vCJD can change depending on the value of life used in calculation and the value of quality of life lost (qaly loss). Table 2 below shows a range of estimates for a case of vCJD based on different values for a statistical life and different values for lost quality of life while suffering from this illness.

Table 2- vCJD Costs per Patient	
Case 1: VSL$5M + $100K*Qaly Loss	
Total medical Costs	$639,782
Total Cost lost utility	$49,285
VSL	$5,000,000
Total Costs Per Case	**$5,689,066**
Case 2: VSL$5M + $300K*Qaly Loss	
Total medical Costs	$639,782
Total Cost lost utility	$147,854
VSL	$5,000,000
Total Costs Per Case	**$5,787,635**
Case 3: VSL$6.5M + $300K*Qaly Loss	
Total medical Costs	$639,782
Total Cost lost utility	$147,854
VSL	$6,500,000
Total Costs Per Case	**$7,287,635**
Case 4: VSL$6.5M + $500K*Qaly Loss	
Total medical Costs	$639,782
Total Cost lost utility	$246,423
VSL	$6,500,000
Total Costs Per Case	**$7,386,205**
Case 5: VSL$5M	
Total medical Costs	$639,782
VSL	$5,000,000
Total Costs Per Case	**$5,639,782**
Case 6: VSL$6.5M	
Total medical Costs	$639,782
VSL	$6,500,000
Total Costs Per Case	**$7,139,782**

The estimated cost per case of variant CJD varies from $5.7 million to $7.1 million depending on the value of a statistical life year (VSLY) and the value of a statistical life (VSL) used. The direct medical costs of care will vary depending on the length of the life of the ill person and the types of doctors seen and the frequency of visits. It is reasonable to assume that if the patient lives for longer than 13 months with the disease, the medical costs and disutility from illness will be higher than stated here.

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